A CASE OF INTRACRANIAL DISEASE INVOLVING THE CHIASM, AND ALSO PRODUCING PROFOUND MENTAL AND NERVOUS DISTURBANCES.*

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Mrs. E. D. T. was first seen by one of the writers (Loveland) on April 30, 1900. She was led into the room apparently totally blind, stooping and tottering as she was helped to a chair. She was of medium height, about thirty years of age, and weighed 140 pounds. Her complexion was slightly pale, but her lips were of good color. Her father was alive and healthy. Her mother died at 48 years, from cancer, which recurred after an operation. As a girl she was healthy, but not robust. The only thing ascertained as illness before marriage, was, that one winter while in school she found it difficult to keep awake, yawned constantly, and could hardly study on account of sleepiness. This condition lasted several weeks.

She was married at 21 years of age, has had two confinements, and one miscarriage, the youngest child being six years old; the miscarriage occurred about seven years ago, midway between the two confinements. Her children were "thin and skinny," as she expresesd it.

She had iritis about a year after marriage, the left eye being affected, but denies ever having had any skin eruption or other signs of specific disease. Some months later I learned that her husband had had a sore which left no scar about a year before his marriage, but denied ever having had any other signs of syphilis. About five years ago there was a period in which she slept very soundly, and did not know on waking what had transpired before she went to sleep, but after her memory was prompted she would recollect. This condition was transient. At this time on one or two occasions she said she could hear, but could not understand what people said to her.

For the past four years she has been well until March 1899, when she ceased to menstruate, and thought herself pregnant, holding that opinion for five months, during which time she ate ravenously hoping for a fat child. On this account she grew very stout. She felt well, and was much occupied with the care of a sick grandmother. At this time she was told that she had a tumor, and must have an operation, but as her mother had died after an operation for cancer, she refused to go to the hospital.

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From the time that she gave up the idea of pregnancy, she slowly lost flesh, but is still fat, while her heaviest weight was 186 pounds. At the same time she began to complain of pain in different parts of her body, and this pain made it difficult for her to walk or assume an erect posture; she slept a great deal, but waked frequently and sat up to ease the pains in her legs. She grew progressively weaker, and until shortly before she came to me (Loveland) was treated for rheumatism.

She had been steadily losing her sight for four months, until now she could scarcely detect light. She complained of great pain in the legs and back, weakness, thirst polyuria, uncontrollable sleepiness, especially in the daytime, and constant yawning when awake. She used to drink a quart or more of water during the night, waking frequently, but going to sleep again after relieving her thirst and emptying her bladder. She passed nearly four quarts of urine in 24 hours, sp. gr. 1006, no albumin or other abnormality.

She never complained of pain in her head or eyes since the iritis mentioned. Her temperature was normal, but her pulse, though regular, was 112. Her tongue was coated and dry. She showed great muscular weakness, and could not make a record on a dynamometer. All her bodily hair had fallen out, and that on her head was dry and falling out. There were no signs of disease of the abdominal or pelvic viscera. There was general hyperesthesia, slightly exaggerated knee-reflexes, and ankle-clonus. There was slight tremor on motion, and a considerable degree of rigidity of muscles, especially of legs and back. She protruded her tongue slowly, and with tremor, but in a straight direction. Her palate was high-arched; pupils were large, and the left one irregular from previous iritis. The pupils reacted feebly to light, but failed to react when all sense of light had been lost a day or two later.

The optic discs were somewhat pale, but beyond this the ophthalmoscope showed nothing to account for her blindness. Retina had also been examined in Feb. 1900, and was again on May 2, by an oculist who was unable to detect any pathological change in the fundus. She seemed very lethargic, or apathetic, both mentally and physically, slow to speak and slow to comprehend. She yawned several times a minute when awake, and would fall asleep while being asked questions.

A diagnosis of specific disease affecting the brain and optic chiasm or tracts was made, and it was thought that some of the symptoms might be hysterical.

She was put on full doses of yellow iodide of mercury, and pills of asafetida and nux vomica.

During the first week or so a test was made almost daily to see if she could detect light, and at her worst she could not locate a Welsbach light a few feet from her, or see bright sunlight. Improvement was rapid under the treatment prescribed, and on May 9 she could detect light but not form, and talked and acted much better. Mercury was discontinued for two or three days on account of signs of salivation, and then given in smaller doses, and strychnia was substituted for the nux and asafetida pill.

On May 12 she could see moving objects, as people or carriages, but could not count fingers. She still complained of pain and soreness, worse in her knees.

On May 14 she saw the colors of a flag, and told what it was, and also walked much better. Read Snellen's 200-ft. test type at 3 1-2 feet.

On May 17 acuity of vision was improved, but hemianopia was noticed. On this date she was taken to Dr. Marlow, and his report of her ocular condition and improvement is as follows:—With the right eye she counted fingers badly at a foot; with the left her vision was 9-140. The pupils reacted promptly to light, but the indirect action of the right was much better than that of the left.

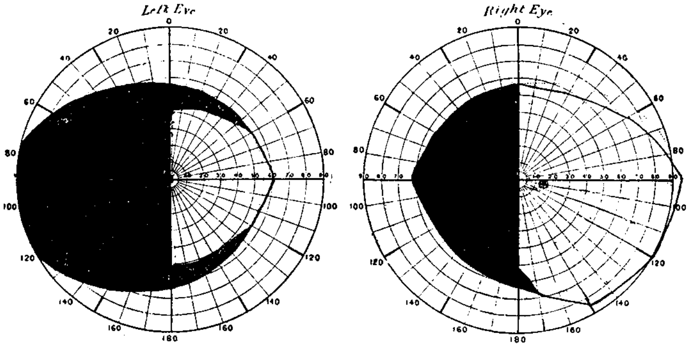
The optic discs were decidedly pale, but there were no white lines along the arteries, or other changes in the blood vessels. The edges of the discs were sharply defined.

Examination of the field of vision showed total loss of the left half of each field, and a partial loss of the right half in the right eye. The condition of the right eye would be better described by saying that vision was present only in a portion of the right half of the field, the fixation point and region around it being included in the blind area. It was, therefore, impossible to make an accurate chart of the field. In the left eye, however, vision was present at the fixation point, consequently the chart of this eye is probably an accurate representation of the field at the time.

On June 1st, vision of the right eye had risen to 6-24, and that of the left to 6-6; the improvement of the conduction of the light impulse in the right eye being shown by the fact that the indirect reaction of the pupil of the left eye had become as good or very nearly as good, as that of the right.

Careful examination of the pupils demonstrated also the presence of the Wernicke hemianopic pupillary inaction sign. Examination of the fields at this time showed marked improvement, mainly in the right eye, and also that the line dividing the seeing from the blind half of the field passed through the fixation point, or within a degree of it. The vision of both eyes improved so that on Aug. 2d it was 6-6 in each eye separately, the left being still a little better than the right.

On Sept. 7th she sees and locates the light of a candle placed near to her in the blind half of the fields, but does not recognize the form of the candle until it crosses into the right half of the field—that is to say, she has a returning light sense in the left half of each field, but no form sense. This change is accompanied by a disappearance of the pupillary hemianopic inaction sign.



Fields showing loss of vision with exception of light sense.

Oct. 13, no change. Letters of the lower line of test type dance or disappear as she looks, large letters do not. As it was thought that this might be due to a rhythmical movement of the eyes necessitated by the fact that the dividing line passed through the fixation point, she was tested on the ophthalmometer, a small printed word being used for fixation, and the corneal image found to be unusually steady, although there was a fine, fairly rhythmical lateral movement.

Nov. 1, tested carefully to determine the gain, and the relation of the dividing line to the fixation point; the result going to show that it lies at a distance of about 5″ from the fixation point. (The button on the perimeter used for fixation measures 5mm. in diameter, and has a screw measuring 3mm. in its center. The white test object is never seen until it impinges on this screw.) This observation was made with great care on each and every occasion upon which I measured the field of vision, with the invariable result of producing a chart of the field in which the line separating the blind from the seeing half passed through the fixation point.

Dec. 13th, color vision normal, but in the right eye there is a relative scotoma for red and green—red being called pink at the fixation point and for about 5° around it; green being called white. In the left eye there is no scotoma.

The history of complete blindness, with dilatation of the pupils, and complete loss of light reflex, without ophthalmoscopic change can only be explained by a lesion lying at the optic commissure, or between it and the corpora geniculata. The fact that the blindness commenced in both eyes, the subsequent history, and the repeated examinations seem to indicate that the lesion, so far as it affected the visual apparatus, probably commenced at the commissure, and gradually receded, leaving the fibers coming from the left optic tract in a practically healthy condition.

The presence of a residual central color scotoma in the right eye, and its entire absence from the left, suggests that the lesion either commenced at, or at some period extended to, the right optic nerve itself, or a part of the chiasm anterior to the point where the crossed and uncrossed fibers join to form the right nerve. For, had it been entirely limited to this point and the parts posterior, we should expect to find the residual color defect hemianopic in character.

In addition to its being an example of that comparatively rare condition—chiasm disease—this case differs from the majority of reported cases of hemianopia chiefly in two respects. First—the hemianopia occurred as a symptom of returning health, rather than of advancing disease. From a condition of complete blindness, we have an almost perfect homonymous hemiopic recovery. Second—the line dividing the seeing from the blind half of the field passes practically through the fixation point. The observations on this point were repeated so many times, and on so many different occasions, that no doubt remained that the test object came well within 15″ and probably to 5″ of the fixation point before it was seen.

It will be remembered that in the vast majority of cases the dividing line does not go through the fixation point, but skirts around it at a variable distance of several degrees, leaving it in the seeing half of the field. So universal is this observation that it has given rise to the theory that the macular region of the retina has a double representation in the cerebral cortex—is, in fact, completely represented in each hemisphere, and consequently in each optic tract.

It is evident that the facts in this case do not harmonize with this theory. From a condition of complete blindness, we have seen recovery of one half of the field with normal central acuity; nevertheless, the recovery in the region of the fixation point, or macula, is strictly hemiopic. We must infer that the optic tract, and the corresponding hemisphere contained fibers from one-half of the macular region only.

It is interesting to note the progress of recovery. From March until the end of April complete blindness, dilatation of pupils, and no reaction to light.

May, latter half, hemiopic restoration of vision, left and right. Reaction of pupils with marked hemianopic pupillary inaction.

June. Rapid improvement in vision.

September shows some return of light sense in the blind (L) halves of the fields, and this is accompanied by the loss of the hemianopic pupillary inaction sign.

October shows improved light sense and projection, but absolute inability to recognize form in left halves of both fields.

From a condition of total blindness, there was a recovery of light, form and color sense on one side, and of light sense on the other half of the field.

The restoration of light sense led to the hope that there might later be a restoration of the form and color sense, but essentially no change has taken place in eight months, so it is unlikely that any further improvement will occur.

During the time covered by Dr. Marlow's report of the patient's eyes, her general condition has improved in a very satisfactory manner. A few things need special note however. On May 19th, saturated sol. iod. potassium, in ten-drop doses, three times a day, was substituted for the yellow iodide of mercury, the doses being increased one drop a day till 25-drop doses were reached. She has taken these doses three weeks out of four up to the present time, the only effect being that she seemed to have a cold in the head when the remedy was first pushed, but this was the only sign of physiological action. Strychnia was also continued for a long time.

As she began to loose her somnolence, and to improve in mental acuity, she seemed to suffer more pain, and slept poorly at night.

In my notes of July 13, 1900, "Can read brevier type at an ordinary distance with either eye, but tires very quickly. Her muscles and eyes both seem very weak, and become exhausted on slight exertion."

September 1. Pain in knees is much better, and she sleeps well at night. She had complained grievously of her knees, but there was never anything objectively wrong with them, except that early in her sickness she could not straighten them, owing to the rigidity of the muscles referred to as one of the symptoms.

November 8. She feels, looks, and acts much better every way, but still is stiff on rising, after occupying any position long. Potassium iodide was ordered to be taken three weeks, then to be omitted for a week. From this date to the present there has been practically no change in her vision, except that she can read or write without such fatigue as she had at an earlier period. She has gained much in both muscular and nervous strength, has done her own housework for a long time, and walks out a good deal. Her grip on the dynamometer only amount to 18, but a year ago she could not make a record on it.

Since this case did not furnish an autopsy, we can only speculate as to the exact location of the lesion in the brain. It was evident at the first that there was some profound cerebral trouble, but it was also plain that the motor tract was at least only indirectly affected; and the somnolence, which suggested the region of intellection, was so like that she had when a girl in school, that it made the family think she was hysterical and indolent, as in the former instance.

The blindness, and later, the ophthalmoscopic examinations rendered us important aid in locating the trouble, viz.: by showing us that optic neuritis was not present, and probably had not existed; that the atrophic appearance of the optic nerve was suggestive of secondary, rather than consecutive atrophy; and consequently we had to deal with a mass pressing on the chiasm and tract, rather than with a basic meningitis.

These considerations, together with the fact that the mental symptoms were more marked than the physical, make it probable that the lesion began in the third ventricle, and that most of the symptoms were caused by pressure; that affecting the visual apparatus beginning at the chiasm or very near to it, and later its influence being limited to the right tract, and that it remained long enough to cause the permanent hemianopic defect. The polyuria and rapid pulse were very slow in passing away. She has not menstruated nor shown signs of a cycle of ovulation.